Abstract:

CONTEXT: Only occasionally, endocrine-active tumors develop directly from hepatic tissue, and may lead to paraneoplastic syndromes (PNS). PNS mostly accompany malignancy of adulthood and are exceedingly rare in children.

PATIENT: A girl aged 6 years and 9 months presented with a 2-month history of rapidly progressive weight gain, abdominal distension, and polyuria/pollakiuria accompanied by short episodes of abdominal pain. She showed the typical clinical features of Cushing's syndrome and a huge hepatic mass. An abdominal computed tomography (CT) scan revealed a large liver tumor. Blood glucose and serum calcium were great elevated.

DESIGN AND OBJECTIVE: Case report describing the causative relationship of the clinical findings.

METHODS: Physical examination; ultrasound of the abdomen; CT scan of the abdomen and the chest; conventional X-rays; routine hematology; blood chemistry and multiple parameters of calcium and phosphorus metabolism; multisteroid analysis in serum and urine; adrenocortical stimulation and suppression tests; histopathological assessment of the resected tumor; immunohistochemistry for ACTH, beta-endorphin, corticotrophin-releasing hormone (CRH), and PTH-related peptide (PTHrP); electron microscopy of tumor cells; ACTH and CRH extraction from the tumor tissue; and clinical follow-up
for more than 20 years. RESULTS: Giant hepatoblastoma (HB; approximately 1000 ml volume) of the right lobe of the liver with combined ectopic ACTH syndrome and PTHrP-induced tumor-associated hypercalcemia. Wide local excision and polychemotherapy led to complete reversal of the paraneoplastic phenotype. CONCLUSIONS: This is the first report of an endocrine-active HB causing both Cushing's syndrome and PTHrP-related 'humoral hypercalcemia of malignancy'. This information should be added to the well-known beta-human chorionic gonadotropin-related paraneoplastic effects of HB in children.